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Case Report

Combined congenital aortic stenosis and coarctation of aorta with severe left ventricle dysfunction and pulmonary hypertension in an infant-one case, multiple challenges

Sourabh Sharma[1], Kritika Sharma[1]*, Sandip Katkade[3]

[1]Department of Cardiothoracic Surgery, Bharat Vikas Parishad Hospital, Kota, Rajasthan, India
[2]Department of Cardiac Anaesthesia, Bharat Vikas Parishad Hospital, Kota, Rajasthan, India
[3]Department of Cardiac Anaesthesia, Sir H. N. Reliance Hospital, Mumbai, Maharashtra, India

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***Correspondence:**
Dr. Kritika Sharma,
E-mail: kriti2289@gmail.com

ABSTRACT

Aortic stenosis (AS) is commonly associated with coarctation of aorta (COA). Due to chronic increase in afterload, it can lead to left ventricle (LV) dysfunction and ultimately LV failure. We present a case of 10-month-old infant with combined AS and COA, complicated by severe LV dysfunction and severe Pulmonary hypertension (PH). There are no guidelines as to which condition should be treated first although both staged and combined procedure have been reported. It poses multiple challenges in the perioperative management. Although performing a balloon aortic valvuloplasty first would reduce the surgical risk for COA repair significantly, but due to lack of facilities for same in this age group, we proceeded with a high-risk COA repair via thoracotomy, avoiding the need for cardiopulmonary bypass (CPB). Application of Aortic cross clamp was associated with a further increase in afterload and hypotension owing to severe LV dysfunction in contrast to the expected increase in proximal pressures, and required high inotrope support to manage hemodynamics. Postoperative course was challenging as well owing to severe pulmonary hypertension. Very few similar cases have been reported in literature.

Keywords: AS, COA, Left ventricle dysfunction, Pulmonary hypertension

INTRODUCTION

Coarctation of aorta (COA) and Aortic Stenosis (AS) both lead to an increased afterload for the left ventricle (LV). A combined lesion with COA and AS would lead to further increase in afterload and rapid progression of LV dysfunction over time. Although very few such cases have been reported in this age, we report the case of an infant with combined COA and AS with severe LV dysfunction which was further complicated by severe pulmonary hypertension.

Ideally, a hybrid or staged procedure with Balloon Aortic Valvuloplasty (BAV) and COA repair should be performed to avoid the risk of complete LV failure due to cross clamp during COA repair. But due to lack of facilities for same and financial constraints, we were able to perform a high-risk COA repair via thoracotomy with inotrope support and minimal cross clamp time. Such a case has rarely been reported and poses multiple challenges for perioperative management which have been discussed in detail.

CASE REPORT

A 10-month-old, 4.5 kg child was brought by the parents with complaints of failure to gain weight and progressive increase in work of breathing. On examination, PR was

132/min, SpO_2 97% and weak femoral pulse. A loud systolic murmur was heard. On further evaluation, the child was diagnosed with severe COA, moderate AS (MG=30), severe tricuspid regurgitation (RVSP 70 mmHg+RAP), with severe left ventricle (LV) systolic dysfunction (LVEF 30%) and severe pulmonary hypertension (PH). The parents were explained in detail about the severity of condition and regarding the high risk associated with surgery and anesthesia.

Due to the complexity of the condition, they were advised to take the child to a higher center where facilities for Balloon aortic valvuloplasty (BAV) were present so a staged or combined hybrid procedure for BAV and COA repair could be planned. Due to financial reasons, the parents refused for same and wished to get operated for COA repair and accepted the risks involved.

After taking consent for high risk for anesthesia and surgery and explaining the risk of perioperative mortality (in view of severe LV dysfunction, severe PH and congenital AS), the child was taken up for COA repair via thoracotomy under general anesthesia (GA).

Adequate blood and blood products were arranged. Inj ketamine 1 mg/kg IV and inj midazolam 0.02 mg/kg was given for premedication and shifted to OT. GA was induced with inj ketamine 2 mg/kg, inj fentanyl 2 mcg/kg and inj glycopyrrolate 10 mcg/kg followed by inj vecuronium 0.1 mg/kg for muscle relaxation. Airway was secured using 4.5 uncuffed ETT. Right radial and left femoral arterial line were secured and a gradient of 20 mmHg was noted between upper and lower limb. 4.5 Fr triple lumen central line was secured in right internal jugular vein.

Surgical end to end anastamosis of aorta was performed via left thoracotomy in 3rd intercostal space (ICS).

Inj. dexamethasone and inj. hydrocortisone were given and inj. mannitol was given for renal protection 15 minutes before the clamp was applied. Once aortic cross clamp was placed, there was a drop in pressures due to severe LV dysfunction and further increase in afterload. CPB pump was primed and kept ready in case of emergency. Inf adrenaline was started at 0.2 mcg/kg/min and Inf dobutamine was started at 5 mcg/kg/min. We were able to maintain MAP of 60-70 mmHg with the support of inotropes in upper limb and MAP 20-30 mmHg in lower limb as shown in Figure 1 where ART is the lower limb pressure and ABP is th upper limb pressure.

Total cross clamp time was 11 minutes. At the time of cross clamp release sodium bicarbonate, vasopressor bolus and fluid bolus were given. After the clamp release, post repair, the upper limb and lower limb pressures almost equalized as shown in Figure 2.

She required high inotrope supports. She was shifted to ICU with inf adrenaline 0.15 mcg/kg/min and Inf dobutamine 5 mcg/kg/min. Urine output was good and hemodynamics were stable.

Figure 1: Hemodynamics after aortic cross clamp was applied where ART is the pressures distal to the clamp (femoral arterial) and ABP is the pressures proximal to the clamp (Right radial).

Figure 2: Hemodynamics post clamp release after repair of coarctation of aorta where upper limb pressure (ABP) and lower limb pressure (ART) are almost equal.

The child was reintubated twice in postoperative period due to repeated episodes of pulmonary hypertension crisis with sudden hypotension, increase in CVP, arryhtmias and desaturation. She was started on inj sildenafil 1 mg/kg/day infusion and inj milrinone 0.5 mcg/kg/min. Other inotropes were tapered and stopped. On POD5, she was successfully extubated with all precautions to avoid PH crisis. Apart from using vasodilators, any external stimulus like suctioning, pain, anxiety was avoided. She was extubated in a deep plane (dexmedetomidine and fentanyl infusion). The rest of

postoperative course was uneventful. On POD7, all dilators were stopped and started on oral sildenafil. She was discharged on POD9 and was advised to undergo BAV in future at a higher center and regular follow up.

DISCUSSION

Left ventricular outflow tract obstruction (LVOTO) is a group of diseases which may be congenital or acquired. It accounts for 4-6% of congenital heart disease (CHD).[1]

The most common congenital LVOTO lesion is congenital AS (70%) followed by COA. Combined AS with COA has an incidence of 5-10% of LVOT obstructive lesions.[2,3]

This group of diseases leads to a chronic increase in afterload, left ventricular hypertrophy, and ultimately LV failure.

This case presented with multiple challenges. Apart from an increase in LV afterload due to both AS and COA, child had severe LV dysfunction which can lead to complete hemodynamic collapse at time of cross clamp for coarctation repair due to sudden, severe increase in afterload. Although, usually cross clamp leads to proximal hypertension and low pressures distal to clamp and requires dilators to control proximal hypertension, a patient with LV dysfunction may not tolerate this increased afterload and can lead to catastrophic acute LV failure.

In adults, with combined AS and COA, a staged or combined hybrid procedure can be planned as there are no guidelines which is better. Also in a staged procedure, if valve replacement is performed first, there is risk of significant hypoperfusion of organs distal to coarctation and if COA repair is planned first, the valvular pathology can cause hemodynamic instability.[4,5]

For congenital AS, BAV or surgical valvotomy has been described in infants with better outcomes in BAV as it avoids the need for sternotomy and CPB.[6] Surgical valvotomy for AS was not advised by us as it would carry high risk owing to poor LV function and small age and need for CPB. Ross procedure which uses native pulmonary valve as an autograft to replace a diseased aortic valve has also been described as a treatment option but not recommended for infants.[7] With regards to timing of Ross procedure, an Italian multicentre registry showed markedly increased mortality of 27.6% for infants as opposed to 0.2% for older children.[8]

For COA, the recommended treatment in small infants is thoracotomy with resection and end to end anastamosis which avoids the need for CPB.[9]

Such a case with combined AS and COA with depressed LV function has rarely been reported.

A similar case with COA and severe LV dysfunction in a 2-month-old child was reported where aortic arch was repaired by median sternotomy under CPB and regional cerebral perfusion and postoperatively child was supported with left ventricle assist device for five days with gradual recovery of left ventricular function.[10]

In our case, the child also had additional AS which would lead to persistent increased afterload after the surgery, with added severe PH.

Due to lack of facilities for mechanical circulatory support and interventional procedures in this age and keeping in mind the financial constraints of family, we decided to go ahead with coarctation repair as a first stage via thoracotomy after discussing in detail with the family about the risks involved and obtaining their consent.

Another interesting single case has been reported where COA repair was performed in an infant using arch to descending thoracic aorta bypass graft which would protect the LV against sudden increase in afterload and also maintain distal perfusion during anastamosis.[9] Although we attempted performing the same technique but it was not possible in our case as the constriction was too proximal and proximal cannula placement in arch was not possible via the thoracotomy incision.

The extubation in postoperative period was extremely challenging and required three attempts owing to severe PH. All necessary precautions were taken in postoperative period to avoid increase in pulmonary artery pressures (PAP). The child was kept sedated; hypoxemia, hypercarbia and acidosis were avoided; any stimulus like pain, anxiety, endotracheal suction were avoided which could cause increase in PAP. Pulmonary vasodilators sildenafil and milrinone were used to control pulmonary hypertension. Extubation was done with ongoing sedation using fentanyl and dexmedetomidine, avoiding any stimulus.

The child was discharged with oral diuretics, sildenafil and digoxin. The LV function improved to 35-40% at the time of discharge.

CONCLUSION

Coarctation of aorta and AS can be complicated with severe LV dysfunction and pulmonary hypertension even in infants. Ideally, a hybrid or staged procedure with BAV and COA repair was required with backup support of mechanical circulatory support in case of complete LV failure due to cross clamp. But due to limited resources and financial constraints, we were able to perform a high risk COA repair via thoracotomy with inotrope support and minimal cross clamp time. Such a case has rarely been reported and poses multiple challenges for perioperative management.

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